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Case report

Sacrococcygeal teratoma with malignant transformation in an adult female: CT and MRI findings

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Abstract. This report describes a case of sacrococcygeal teratoma with adenocarcinomatous transformation in a 45-year-old woman. This is an infrequent location for teratoma in adults and malignant transformation has rarely been described. Prognosis depends on complete excision. Clinical manifestations, imaging aspects and histological findings of this case are presented. CT and MRI adequately document the mixed cystic and solid nature of the tumour, its extension and relations with adjacent structures, allowing accurate pre-operative planning.

Sacrococcygeal teratoma is the most common solid neoplasm in neonates, with an estimated prevalence of 1 in 35 000–40 000 live births. They can be diagnosed prenatally by fetal ultrasound and 50–70% are found during the first few days of life. 80% are diagnosed by the sixth month and fewer than 10% beyond the age of 2 years [1, 2]. Reported cases of sacrococcygeal teratomas in adults are rare. Reviews of the literature by Head et al [3], Ahmed and Pollock [4], Ng et al [5] and Bull et al [6] found a total of 88 reported cases, with only 16 described as malignant or with malignant transformation. Treatment consists of complete surgical resection. Long-term survival is possible for malignant tumours, but incomplete surgical removal carries a poor prognosis. If the tumour invades adjacent structures, neoadjuvant pre-operative combination chemotherapy should be given [7].

We report an unusual case of a 45-year-old woman with intestinal and urinary obstructive symptoms due to a large sacrococcygeal teratoma with adenocarcinomatous transformation. CT and MRI appropriately demonstrated the combined cystic and solid nature of the tumour, its extension and relations with adjacent structures, allowing accurate pre-operative planning.

Case report

A 45-year-old woman with a 1-year history of gradually increasing lower back and pelvic pain, presented with constipation, dysuria and urinary frequency that had progressively developed over 2 months. There was an obstetrical history of dystocic forceps deliveries at ages 21 and 27. On gynaecological and rectal examination a large palpable mass was found in the right pelvis, posterior to the rectum. Neurological examination was unremarkable. Alpha-fetoprotein, carcinoembryonic antigen and human chorionic gonadotropin levels were normal. On pelvic CT (Figure 1) a regularly marginated thin walled cystic mass measuring 15 cm × 11.5 cm × 10.5 cm was identified, anterior to the sacrum and coccyx and extending inferiorly into the left ischiorectal fossa, compressing the urinary bladder and sigmoid colon and anteriorly displacing the rectum and vagina. Its content had homogeneous density near to that of water (10 Hounsfield Units), with a discrete amount of floating fat. At the coccyx bone level, a contrast enhancing 2.5 cm solid nodule was also evident as vegetation from the posterior wall. No evidence of bone destruction or invasion of the adjacent structures was found. Hysterectomy, bilateral adnexectomy and biopsies of the tumour were performed at laparotomy. Histopathological evaluation showed the wall of the cystic portion to be composed of a fibrous capsule with an inner lining of respiratory epithelium and areas of squamous epithelium. The described solid nodule was an adenocarcinoma. These findings were concordant with malignant transformation in a sacrococcygeal teratoma, and the patient was referred to our institution for further evaluation and treatment.

Subsequent MRI, for better pre-excisional evaluation, showed two distinct, well circumscribed, thin walled cystic components with

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(a) (b) (c)

Figure 1. CT scans show (a) a thin walled cystic mass with homogeneous hypodense content anterior to the sacrum, compressing the urinary bladder and anteriorly displacing the rectum and vagina. A discrete amount of floating fat can be seen anteriorly (arrowhead). (b) At an inferior level a contrast enhancing solid nodule is seen, apparently vegetating from the posterior wall, in close relation with the coccyx. There is no evidence of bone destruction. (c) The mass extends inferiorly into the left ischiorectal fossa.

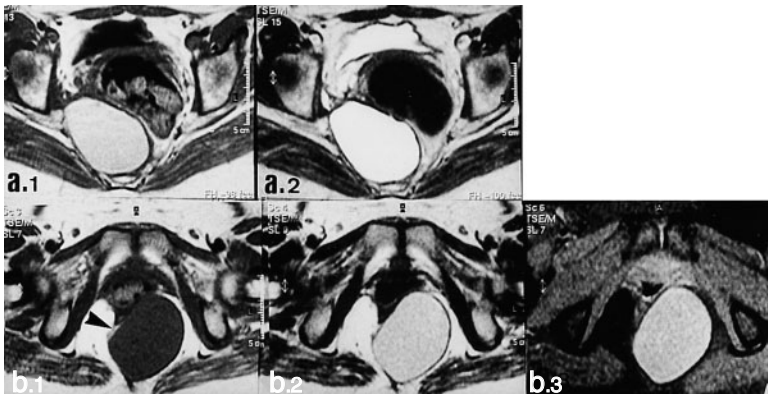

Figure 2. MRI reveals two independent cystic portions. Axial scans show (a) a right-sided superior and (b) a left-sided inferior cyst with homogeneous contents. The superior cyst shows hyperintense signal on both (a.1) T_1 and (a.2) T_2 weighted images, compatible with high protein fluid. The inferior cyst shows (b.1) hypointense signal on T_1 and (b.2) hyperintense signal on T_2 weighted images, indicating serous fluid. (b.1) Only the discrete lamella of hyperintense signal, visible at the right periphery of the inferior cyst (arrowhead) corresponds to pure fat content, as it is completely suppressed on a fat saturated sequence (b.3). a.1 and b.1, T_1 weighted images (TR, 426 ms; TE, 8.0 ms); a.2 and b.2, T_2 weighted turbo spin echo images (TR, 5404 ms; TE, 120 ms); b.3, fat saturated T_1 weighted image (TR, 680 ms; TE, 8.0 ms).

homogeneous high protein and serous fluid contents, respectively (Figure 2). There was a discrete lamella of fat at the right periphery of the inferior cyst. Separating the cysts, a mass corresponding to the solid nodule on CT was evident after administration of gadolinium (Figure 3). Coronal and sagittal planes (Figure 4) displayed the tumour's hourglass shape, with the rectosigmoidal transition seen tightly compressed between the two cystic components. Midline sagittal images clearly illustrated the solid components close relation with the anterior surface of

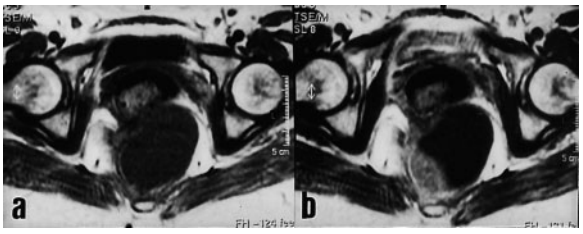

Figure 3. Axial T_1 weighted (TR, 426 ms; TE, 8.0 ms) scan. (a) Without contrast the adenocarcinomatous nodule has hypointense signal, indistinguishable from the content of the inferior cyst. (b) After gadolinium administration the nodule becomes distinctively hyperintense.

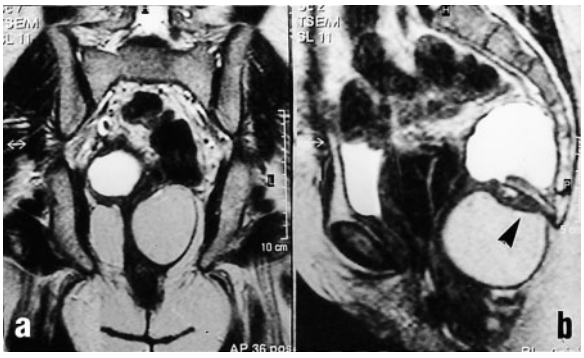

Figure 4. (a) Coronal and (b) midline sagittal T_2 weighted turbo spin echo MR images (TR, 6859 ms; TE, 150 ms). The tumour has an hourglass appearance with the rectosigmoidal transition tightly compressed between the cystic components. The solid component at the isthmus (arrowhead) is seen in close relation with the anterior surface of the coccyx. No evidence of invasion of the surrounding structures was seen in any of the planes.

the coccyx. As on CT, there was no evidence of invasion of the rectal or bladder wall, surrounding fat, pelvic floor muscles or osseous structures in any of the planes, nor was regional lymph node enlargement seen.

The patient was re-operated on, with complete excision of the tumour. On gross examination the surgical specimen looked like a biloculated cyst filled with sebum, haemorrhagic material of a necrotic appearance and serous fluid, separated by a firm heterogeneous solid nodule.

Microscopically, the wall of the cyst was lined with mature stratified squamous keratinizing epithelium. Areas of respiratory epithelium were also present. The solid region was composed of irregular nests of neoplastic glands with morphological features of a moderately differentiated adenocarcinoma, infiltrating the surrounding stroma with desmoplastic features (Figure 5), hence confirming the diagnosis of adenocarcinomatous transformation in a mature teratoma. Soft tissue margins were negative for tumour, which was found confined to the limits of the surgical specimen. No angioinvasion was identified.

The patient underwent post-operative adjuvant combination chemotherapy with three cycles of bleomicin (30 mg, day 1 and day 8), etoposide (100 mg m^{-2}, days 1–5) and cisplatinum (20 mg m^{-2}, days 1–5). There was no evidence of recurrence by the twelfth month of follow-up.

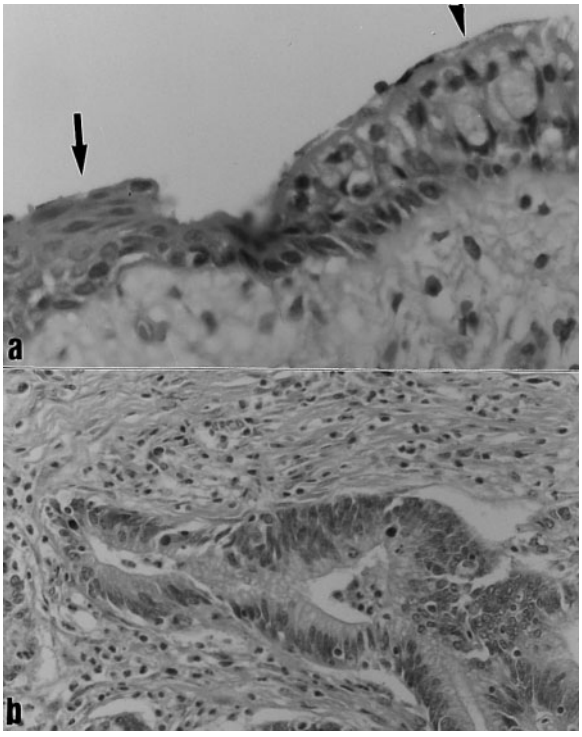

Figure 5. Photomicrographs (haematoxylin-eosin stain, original magnification (a) ×40; (b) ×20) show (a) the wall of the cyst composed of fibrous tissue with an inner lining of mature squamous epithelium and respiratory epithelium, and (b) irregular nests of moderately differentiated adenocarcinomatous glands infiltrating the surrounding stroma.

Discussion

Teratomas are composed of tissues that are strange to the anatomic site of appearance and disposed in a disorganized fashion. They derive from embryonic pluripotent cells and may have various degrees of maturation, according to which they are classified as mature, immature, and malignant (malignant tissue of germ cell origin) [1, 2]. Teratomas with malignant transformation contain malignant cells derived from mature tissues. Pluripotent cells are normally present in the gonads, and may also be found in abnormal sequestered midline embryonic rests. Accordingly, teratomas are found with decreasing frequency in the ovaries and testis, mediastinum, retroperitoneal space, sacrococcygeal zone, pineal and other intracranial locations [5]. Sacrococcygeal teratomas are thought to originate from multipotential cells in Henson's node, which migrates caudally to rest in the coccyx [8]. They may grow posteroinferiorly into the gluteal area and/or anterosuperiorly into the abdominopelvic cavity [9]. There is a tendency among the paediatric population toward malignant transformation of sacrococcygeal teratomas with increasing age. However, in case reports concerning adult patients, benign tumours predominate [5]. Exclusively presacral tumours present later than those with an external component, and have a higher prevalence of malignant transformation. They can present with chronic fistula, low back pain or obstructive symptoms of the gastrointestinal or genitourinary tracts. In the reported case the sacrococcygeal teratoma might have been the cause of the dystocic forceps deliveries. Conventional studies performed with contrast material may demonstrate extrinsic narrowing with anterior displacement of the rectum, lateral displacement of the rectosigmoid colon and compression of the urinary bladder against the anterior abdominal wall, caused by a space occupying presacral lesion. CT and MRI are the most important investigations for characterization of the mass, evaluation of its intrapelvic extension and relationship to other structures. Most commonly, teratomas appear as a complex mass with roughly equal amounts of solid heterogeneous and cystic areas with or without septations. They also frequently present as thick walled cystic masses, sometimes multiloculated, that may contain fat, calcified elements and/or small solid nodules, as in the reported case. Predominantly solid masses are uncommon [2]. The reported case, with two cystic components, a solid nodule and a small amount of fat, was suggestive of the second

described pattern of teratoma. Complex, predominantly solid tumours are more likely to be malignant and significant areas of necrosis within the tumour, or poor definition of adjacent soft-tissue planes, are signs suggestive of malignancy. Invasion of adjacent structures, rather than simple displacement, sacral destruction and secondary findings such as locoregional lymph node and distant metastases are clearly indicative of malignancy [1, 2]. Nevertheless, imaging features alone do not allow definite differentiation between benign teratomas and those with malignant transformation. In the absence of aspects suggestive of malignancy, benignity must not be assumed.

Differential diagnosis of sacrococcygeal masses varies with their nature. In the adult, presacrococcygeal simple cystic lesions may correspond to anterior meningocele, rectal or anal duplication cyst or anal gland cyst. In the appropriate clinical context they may also represent abnormal collections like seroma or urinoma. In the presence of a multiloculated cystic lesion, a tail gut cyst (retrorectal cystic hamartoma) must be considered. Denser and more complex lesions may represent chronic retrorectal abscess, pilonidal or dermoid cyst, soft tissue or bone tumours. With destructive or invasive imaging features, a malignant nature must be considered including sarcomatous tumours, chordoma or metastases. Osteomyelitis of the sacrum may also cause osseous destruction with associated soft tissue mass. Sacrococcygeal teratoma should be included in the differential diagnosis, whatever the nature of the lesion [1, 7].

CT is the most sensitive method of demonstrating calcification/ossification, which may be visible in over 50% of malignant tumours [1], and the integrity of adjacent cortical bone. Fat–fluid or fluid–debris levels may appear with complex cyst contents. MRI better depicts cystic elements, the contents of which might be inferred from signal intensity patterns on different sequences. Fat components can be identified by either CT or MRI. CT and MRI are complementary in the evaluation of sacrococcygeal column anomalies such as spinal dysraphism, sacral agenesis, pressure erosion and remodelling that can be associated with sacrococcygeal teratomas.

The treatment for all sacrococcygeal teratomas consists of early and complete surgical excision with coccygectomy. This bone may contain a nidus of pluripotent cells that increase recurrence rates to 37% when not excised [1, 5, 10]. Surgical approach depends on size and topographic location of the tumour [10–13]. Owing to its direct multiplanar imaging capacities, MRI allows a better topographic evaluation of the tumour that is important for accurate pre-operative planning. Solid teratomas may be very vascular, causing important intraoperative haemorrhage [11, 14]. Pre-operative angiography may be considered for previous blood supply evaluation and embolisation in larger tumours [1, 8]. If complete resection is accomplished, benign teratomas have a good prognosis and long-term survival is possible with malignant tumours. Because malignant transformation is rare there has been no standard recommendation for the use of chemotherapy or radiation and the best treatment plan seems to be referral to an oncology centre, where individualized multimodality therapy can be achieved [6, 12].

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